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Vertigo in Children

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Academic dissertation
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on 15 September 2006, at 12 noon.

Helsinki 2006

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ISBN 952-92-0817-0 (paperback)
ISBN 952-10-3332-0 (PDF), available at http://ethesis.helsinki.fi

University Printing House
Helsinki 2006

To Janne, Arttu, Jaakko,
Fanny, and Sandra

Abstract

Vertigo in children is more common than previously thought. However, only a small fraction of affected children meet a physician. The reason for this may be the benign course of vertigo in children. Most childhood vertigo is self-limiting, and the provoking factor can often be identified.

The differential diagnostic process in children with vertigo is extensive and quite challenging even for otologists and child neurologists, who are the key persons involved in treating vertiginous children. The cause of vertigo can vary from orthostatic hypotension to a brain tumor, and thus, a structured approach is essential in avoiding unnecessary examinations and achieving a diagnosis. Common forms of vertigo in children are otitis media-related dizziness, benign paroxysmal vertigo of childhood, migraine-associated dizziness, and vestibular neuronitis. Orthostatic hypotension, which is not a true vertigo, is the predominant type of dizziness in children. Vertigo is often divided according to origin into peripheral and central types. An otologist is familiar with peripheral causes, while a neurologist treats central causes. Close cooperation between different specialists is essential. Sometimes consultation with a psychiatrist or an ophthalmologist can lead to the correct diagnosis.

The purpose of this study was to evaluate the prevalence and clinical characteristics of vertigo in children. We prospectively collected general population-based data from three schools and one child welfare clinic located close to Helsinki University Central Hospital. A simple questionnaire with mostly closed questions was given to 300 consecutive children visiting the welfare clinic. At the schools, entire classes that fit the desired age groups received the questionnaire. Of the 1050 children who received the questionnaire, 938 (473 girls, 465 boys) returned it, the response rate thus being 89% (I). In Study II, we evaluated the 24 vertiginous children (15 girls, 9 boys) with true vertigo and 12 healthy age- and gender-matched controls. A detailed medical history was obtained using a structured approach, and an otoneurologic examination, including audiogram, electronystagmography, and tympanometry, was performed at the Helsinki University Central Hospital ear, nose, and throat clinic for cooperative subjects. In Study III, we reviewed and evaluated the medical records of 119 children (63 girls, 56 boys) aged 0-17 years who had visited the ear, nose, and throat clinic with a primary complaint of vertigo in 2000-2004. We also wanted information about indications for imaging of the head in vertiginous children. To this end, we reviewed the medical records of 978 children who had undergone imaging of the head for various indications. Of these, 87 children aged 0-16 years were imaged because of vertigo. Subjects of interest were the 23 vertiginous children with an acute deviant finding in magnetic resonance images or computerized tomography (IV).

Our results indicate that vertigo and other balance problems in children are quite common. Of the Helsinki University Central Hospital area population, 8% of the children had sometimes experienced vertigo, dizziness, or balance problems. Of these, 23% had vertigo sufficiently severe to stop their activity (I). The most common forms of vertigo in the children examined at the clinic were otitis media-related vertigo, benign paroxysmal vertigo of childhood, and migraine-associated dizziness. More headaches and head traumas were observed in vertiginous children than in healthy controls (II). The most common di-

agnoses of clinic patients within the five-year period were benign paroxysmal vertigo of childhood, migraine-associated dizziness, vestibular neuronitis, and otitis media-related vertigo. Valuable diagnostic tools in the diagnostic process were patient history and oto-neurologic examinations, including audiogram, electronystagmography, and tympanometry (III). If the vertiginous child had neurological deficits, persistent headache, or preceding head trauma, imaging of the head was indicated (IV).

Contents

Abbreviations

Ag..... audiogram
BPVoC ..... benign paroxysmal vertigo of childhood
CT ..... computerized tomography
EEG..... electroencephalography
ENG ..... electronystagmography
ENT..... ear, nose, and throat
HUCH ..... Helsinki University Central Hospital
MAD ..... migraine-associated dizziness
MRI ..... magnetic resonance imaging
OME..... otitis media with effusion
OM ..... otitis media
PLF..... perilymphatic fistula
SNHL ..... sensorineural hearing loss
VN..... vestibular neuronitis

List of original publications

This thesis is based on the following original publications, referred to in the text by Roman numerals I-IV:

I Niemensivu R, Pyykkö I, Wiener-Vacher S, Kentala E. Vertigo and balance problems in children- an epidemiologic study in Finland. Int J Pediatr Otorhinolaryngol 70(2): 259-65, 2006.

II Niemensivu R, Kentala E, Wiener-Vacher S, Pyykkö I. Evaluation of vertiginous children. (submitted).

III Niemensivu R, Pyykkö I, Kentala E. Vertigo and imbalance in children: a retrospective study in a Helsinki University Otorhinolaryngology clinic. Arch Otolaryngol Head Neck Surg 131(11): 996-1000, 2005.

IV Niemensivu R, Pyykkö I, Valanne L, Kentala E. Value of imaging studies in vertiginous children. Int J Pediatr Otorhinolaryngol 70: 1639-44, 2006.

1 Introduction

Vertigo in children is infrequent and when it manifests it has a different clinical picture than in adults. In the literature, childhood vertigo has received much less attention than vertigo occurring in adulthood. Among otologists and child neurologists, the key clinicians treating vertiginous children, the differential diagnosis is not well established. Vertigo in children is a diagnostic challenge for clinicians because of their immature peripheral and central vestibular systems and limited communication abilities (Eviatar and Eviatar 1977; Balkany and Finkel 1986). Meniere's syndrome is rare in children, while benign paroxysmal vertigo of childhood (BPVoC) and other migraine equivalents are more common than in adults. Benign paroxysmal positional vertigo occurs very seldom in children because cupular deposits are a phenomenon of the aging vestibular labyrinth (Bachor et al 2002). Brainstem and cerebellar tumors are relatively more common in children than in adults (Britton and Block 1988). The prevalence of vertigo in children remains unknown.

Vertigo in children, as in adults, is often divided into peripheral and central causes. Peripheral causes include otitis media (OM)-related vertigo, BPVoC, Meniere's disease (MD), posttraumatic vertigo, perilymphatic fistula (PLF), vestibular neuronitis (VN), and labyrinthitis (Blayney and Colman 1984; Balkany and Finkel 1986). Cochlear symptoms may be associated with vertigo or vertigo can occur as an isolated symptom, as in BPVoC, VN, and vertigo caused by vestibulotoxic drugs (D'Agostino et al. 1997). The most common central causes of vertigo or dizziness are epilepsy, migraine, multiple sclerosis, and tumors of the central nervous system (CNS). The predominant forms of vertigo in children are OM-related vertigo and BPVoC (Blayney and Colman 1984; Bower and Cotton 1995), followed by migraine-associated dizziness (MAD) (Bower and Cotton 1995; Ravid et al. 2003), and vertigo of unknown origin (Blayney and Colman 1984). In one study, cranial trauma was a leading cause of vertigo in children, followed by BPVoC, which is diagnosed by typical clinical symptoms and exclusion of all other known forms of vertigo (D'Agastino et al. 1997). When children with OM were excluded in another study, MAD, BPVoC, posttraumatic vertigo after head trauma, and MD were the prevailing forms of vertigo (Choung et al. 2003). Orthostatic hypotension is a very common physiological phenomenon in children, although it is not considered to be true vertigo.

Because many possible etiologies exist for vertigo, a systemic and structured approach that takes into account the patient's age and the complaint is essential. Several authors offer algorithms to facilitate the evaluation process and eliminate unnecessary and expensive examinations (Blayney and Colman 1984; Eviatar 1994; Ravid et al. 2003). Evaluation of a vertiginous child requires close cooperation between different specialists, with the most important roles often being filled by otologists (Britton and Block 1988) and neurologists (Blayney and Colman 1984; Balkany and Finkel 1986).

A thorough history is the most valuable diagnostic tool in the evaluation process. A physical examination, particularly an otoneurologic examination, including audiometry, tympanometry, and electronystagmography (ENG), and in selected cases imaging of the head, laboratory tests, and electroencephalography (EEG) help to confirm the diagnosis (Balkany and Finkel 1986; Eviatar 1994; Bower and Cotton 1995; Ravid et al. 2003).

The aim of this work was to evaluate the prevalence of vertigo in children, the etiological factors causing vertigo, and the methods employed to make the diagnosis. A further objective was to determine the value of head imaging in vertiginous children.

2 Review of the literature

2.1 History

The word vertigo derives from the Latin verb vertere (to turn). Dizziness, in turn comes from the old English word dysig (stupid). The etymology of ataxia originates from the Greek verb tassein (to put in order) (Merriam-Webster online dictionary).

The Columbia Encyclopedia Dictionary gives the following definition of vertigo: "Sensations of moving in space or of objects moving about a person and the resultant difficulty in maintaining equilibrium. True vertigo, as distinguished from faintness, lightheadedness, and other forms of dizziness, occurs as a result of disturbance of some part of the body's balancing mechanism, located in the inner ear (e.g. vestibule, semicircular canals, auditory nerves). Labyrinthitis, or infection and irritation of the middle and inner ear, is a common cause of vertigo. Elimination of infectious, toxic, or environmental factors underlying the disturbance is essential for permanent relief " (Columbia Encyclopedia Dictionary, sixth edition 2001-2005). Vertigo is defined as an illusion of motion of the patient or of his/her environment. Vertigo implies a true equilibrium disturbance and is often caused by problems in the inner ear balance organ (Eviatar 1994; Ravid et al. 2003).

For the term dizziness, the same dictionary refers the reader to vertigo. Dizziness is also described by the Merriam-Webster online dictionary as having a whirling sensation in the head with a tendency to fall or of being mentally confused. Dizziness is a nonspecific complaint that can describe many sensations, including lightheadedness, imbalance, or disequilibrium. It can be the manifestation of such psychological disorders as panic attacks or depression, or orthostatic hypotension (Eviatar 1994; Ravid et al. 2003). Dizziness can also be described as a disturbed sense of relationship to space, or unsteadiness with a feeling of movement (Bower and Cotton 1995).

Ataxia is described as lack of coordination of the voluntary muscles, resulting in irregular movements of the body. Ataxia can be brought on by an injury, infection, or a degenerative disease of the CNS, e.g. syphilis, encephalitis, brain tumor, or multiple sclerosis. (Columbia Encyclopedia Dictionary, sixth edition 2001-2005). In the Finnish language, the word "huimaus" is often used to describe both vertigo and dizziness, sometimes also ataxia.

2.2 Prevalence of vertigo in childhood

2.2.1 Population-based studies

Only one epidemiological study based on the general population of 2165 children is available. This Scottish study notes that vertigo in children is common but seldom diagnosed. The prevalence of vertigo was 14%. The authors defined paroxysmal vertigo as "at least three transient episodes of vertigo of the child or of the environment, severe enough to interfere with normal activities and not associated with loss of consciousness or neurological deficits". Altogether 2% of children fulfilled these criteria (Russell and Abu-Arafeh 1999).

2.2.2 Hospital-based studies

The prevalence of vertigo in children varies considerably according to the specialization of the clinic where the study is done (Table 1). The most common forms of vertigo in the field of otology are as follows: OM-related dizziness, BPVoC, and unknown etiology (Blayney and Colman 1984; Bower and Cotton 1995), labyrinthitis (Blayney and Colman 1984), and posttraumatic vertigo after head trauma (D'Agostino et al.1997). In neurological studies, epilepsy-related vertigo (Eviatar and Eviatar 1977), MAD, BPVoC, and psychic origin vertigo were predominant forms of vertigo (Eviatar and Eviatar 1977; Weisleder and Fife 2001; Ravid et al. 2003). VN was also frequently seen in neurology clinics (Eviatar and Eviatar 1977; Ravid et al. 2003).

2.3 Vertigo diseases in children

In studies based on examinations of patients at otolaryngology clinics, a higher incidence of peripheral causes of vertigo, such as OM-related vertigo and VN, was seen than in studies in which patients have been referred to neurology clinics (Table 1). The most common forms of vertigo in children are described in the sections below.

Table 1: Prevalence of most common causes of vertigo in selected studies

Study	Bower and Cotton 1995	Blayney and Colman 1984	D'Agostino et al. 1997	Choung et al. 2003	Eviatar and Eviatar 1977	Weisleder and Fife 2001	Ravid et al. 2003
Number of children	34	27	282	55	50	31	62
Clinic	ENT	ENT	ENT	ENT*	Neurology	Neurology	Neurology
Peripheral causes							
Otitis media	21 %	19 %		*			
BPVoC	15 %	18 %	21 %	25 %	4 %	19 %	16 %
Vestibular neuronitis	9 %		1 %	2 %	10 %		14 %
Meniere's disease	6 %			4 %		6 %	
Endolymphatic hydrops				4 %			
Perilymphatic fistula		4 %	1 %				
Sudden hearing loss						3 %	
BPPV				4 %			
Labyrinthitis	3 %	11 %	18 %				
Peripheral total	54 %	52 %	41 %	39 %	14 %	28 %	30 %
Central causes							
MAD	12 %	7 %	5 %	31 %	10 %	35 %	39 %
Head trauma	9 %	4 %	30 %	7 %	8 %		3 %
Epilepsy		4 %	3 %	2 %	50 %		3 %
Infection (CNS)					6 %		
Demyelinating disease			2 %				
Central origin lesions	3 %	4 %	9 %	2 %			
Central total	24 %	19 %	49 %	42 %	74 %	35 %	45 %
Other							
Psychic					10 %	10 %	13 %
Unknown	12 %	30 %	9 %	18 %		19 %	
Familial ataxia	3 %					3 %	
Orthostatic hypotension							9 %
Others total	15 %	30 %	9 %	18 %	10 %	32 %	22 %

BPVoC, Benign paroxysmal vertigo of childhood
BPPV, Benign paroxysmal positional vertigo
MAD, Migraine-associated dizziness
CNS, Central nervous system
ENT, Ear, nose, and throat

* children with abnormal eardrums or tympanograms were excluded

2.3.1 Migraine variants and complicated migraine

Migraine headaches and their variants, although rare, are the most common episodic disorders in children. The neurological disturbances can sometimes mimic strokes, seizures, movement disorders, and other diseases. These manifestations of migraine in childhood are called complicated migraine or migraine variants. Childhood migraine variants may be precursors to or associated with migraine (Parker 1997). The five known migraine variants (also known as migraine-equivalent syndromes), which exist with or without headache, are BPVoC, cyclic vomiting, infantile torticollis, acephalic migraine, and acute confusional migraine (Parker 1997; Al-Twaijri and Shevell 2002).

Complicated migraine headaches are unusual neurological symptoms that occur during the course of a migraine headache. Headache is almost always present, but because of the child's disorientation, it cannot always be related to migraine. Ophthalmoplegic migraine, retinal migraine, hemiplegic migraine, basilar artery migraine, and acute confusional migraine are forms of complicated migraines. Patients with complicated migraine are at higher risk for having strokes (Parker 1997).

Children with infantile torticollis are infants or toddlers. The attacks last from hours to days and are often associated with nausea, vomiting, pallor, and agitation. Older children with infantile torticollis may have ataxia or vertigo as well (Parker 1989; Parker 1997). The age of onset of BPVoC and cyclic vomiting varies markedly (from 9 months to 13 years in both), with the mean age of onset being 5 and 6.5 years, respectively, whereas acephalic migraines and acute confusional migraines are largely disorders of mid-childhood to adolescence (Al-Twaijri and Shevell 2002). If the episode in cyclic vomiting is prolonged, it leads to dehydration (Parker 1997). In cyclic vomiting and acephalic migraine, coexisting typical migraine syndromes were observed in more than half of cases. There is a clear gender predominance for females and a strong history of migraine in all subtypes, ranging from 65% (cyclic vomiting) to 100% (acute confusional migraines) (Al-Twaijri and Shevell 2002).

Several features distinguish seizures and complicated migraines. A patient with complicated migraine often has a history of typical migraine attacks, the evolution of symptoms and signs is slower, the patient during and after the migraine attack more often has a memory of the event, and nausea and vomiting are more common (Parker 1997). Basilar artery migraine syndrome, which is a form of complicated migraine, occurs primarily in teenage girls, but may begin earlier in childhood (Parker 1997). Vertigo is a relatively common symptom in basilar artery migraine (Parker 1989).

Episodic ataxia type 2 (EA2) is an autosomal dominant episodic neurologic syndrome characterized by hours-long paroxysmal ataxia, attacks of vertigo, fluctuating generalized weakness, nausea, vomiting, and nystagmus. Migraine headaches occur in more than half of the genetically defined patients. EA2 episodes typically begin before the age of 20. The episodes are triggered by stress and exertion. A wide range of mutations in the CACNA1A gene are associated with EA2. Low total cerebellar creatine can be an early sign of calcium channel dysfunction in EA2 patients (Jen et al. 2004; Harno et al. 2005).

2.3.2 Benign paroxysmal vertigo of childhood

In 1964, Basser first described BPVoC and its typical clinical features. In his study, he also considered differential diagnostics of childhood vertigo, but did not associate BPVoC with migraine. While no proven etiology exists, BPVoC is considered to be a migraine variant, equivalent, or precursor (Finkelhor and Harker 1987; Parker 1989; Lanzi et al. 1994; Parker 1997; Russell and Abu-Arafeh 1999; Drigo et al. 2001; Al-Twaijri and Shevell 2002;). A family history of migraine can almost always be obtained, and the child later develops more typical migraine attacks (Parker 1997).

BPVoC is a vestibular disorder characterized by sudden brief episodes of spinning vertigo, rarely lasting more than a few minutes. During the attack the child is conscious and often frightened, afterwards continuing to play as though nothing had happened. Pallor is frequently associated with the attack, and nystagmus, sweating, and vomiting are occasionally present. The attacks may occur in any position and are not provoked by head posture or movement. The child typically clutches an adult until the attack ceases. The attacks are recurrent and may occur from many times per day to a few times per month. Typical age of onset is the first four years of life, but 5-10 years is also possible. Soon after onset, the attacks become more frequent, then gradually decreasing and disappearing around the age of 10 years (Finkelhor and

Harker 1987). Cass et al. (1997) reported that children with BPVoC did not necessarily report true vertigo, but more commonly described imbalance, movement-associated disequilibrium, or paroxysmal lightheadedness.

The diagnosis of BPVoC is based on a typical clinical picture because all radiological evaluations and otologic and neurological examinations, including ENG, audiogram (Ag), and EEG, are normal. There is no treatment for BPVoC, and the attacks are self-limiting (Finkelhor and Harker 1987; Parker 1997). Unrecognized, BPVoC can worry parents and be a discomfort for the child. It is important to inform the family of the benign course of the disorder. BPVoC is usually diagnosed only after the exclusion of all other known forms of vertigo (D'Agastino et al. 1997).

2.3.3 Migraine-associated dizziness

Migraine has long been associated with vertigo, but controlled studies in children are lacking. In a study of adults, vertigo occurred with or without headache and the duration of attacks varied from minutes to days (Neuhauser et al. 2001).

Migraine headaches and their variants are the most common recurrent episodic disorders in children. About 5-10% of children suffer from migraines (Parker 1997). Migraines become more common as the child gets older, but this increase may simply be due to underdiagnosis of migraine in younger children. There is a strong positive family history of migraine (Parker 1997). A typical child with vestibular migraine is a teenage girl with recurrent episodes of headache and dizziness, a past history of motion sickness, a family history of severe headaches, and a normal neurological examination (Weisleder and Fife 2001). The current International Headache Society (IHS) classification of migraine does not include vertigo as a symptom, although an association exists between migraine and vertigo and dizziness. This association can be subdivided into causal, statistical, and coincidental associations (Neuhauser and Lempert 2004).

The criteria for MAD (known also in literature as migrainous vertigo) are the following: recurrent paroxysmal vertigo attacks, current or previous history of migraine (IHS criteria), at least one migraine symptom (headache, phono- or photophobia, visual or other auras), and at least two separate vertigo attacks, with other causes excluded. The pathophysiology of MAD is unclear (Neuhauser and Lempert 2004).

MAD is important in the differential diagnosis of vertigo with spontaneous and positional nystagmus. MAD can present in both central and peripheral vestibular disorders (von Brevern et al. 2005). Vestibular symptoms with migraine are common, and vestibular and auditory deficits may be temporary or permanent in children and adults (Harker and Rassekh 1987). Migraine-associated vestibular symptoms can occur prior to the onset of headache, during headache, or without headache (Parker 1989; Cass et al. 1997). Headache is considered to be migrainous if it is described as pulsative or throbbing, localized, hemicranial, lasting for several hours, and occurring in conjunction with nausea or vomiting and phono- and photophobia.

The diagnosis of migraine-associated vestibulopathy requires awareness and a careful history-taking, including family history (Harker and Rassekh 1987; Parker 1989; Cass et al. 1997; Weisleder and Fife 2001). No specific biological markers exist for MAD, and it is diagnosed on the basis of history (Neuhauser and Lempert 2004). Correct diagnosis is important to ensure

optimal treatment and to avoid unnecessary examinations or surgery (Harker and Rassekh 1987). The diagnosis of juvenile migraine is purely clinical, and a family history, primarily maternal, supports the diagnosis. The most common early symptoms consist of vomiting, behavioral changes, sleep problems, pallor, vertigo or ataxia, and headaches (Barlow 1994).

Brain imaging is indicated for patients whose symptoms deviate from the typical migraine profile (Weisleder and Fife 2001). A trial of prophylactic migraine medication, especially when episodes are frequent and disabling, should be considered for both diagnostic and therapeutic purposes (Weisleder and Fife 2001). Treatment strategies for MAD include avoidance of stress and dietary triggers, such as tyramine-containing foods, alcohol, and caffeine, and adequate sleep and rest. The underlying migraine should be treated with medicine and possibly anti-motion sickness medication. An associated anxiety or panic disorder should be treated with behavioral therapy or pharmacotherapy, or both (Cass et al. 1997).

2.3.4 Otitis media-related vertigo

OM is the leading cause of healthcare visits by children, and it is the prevailing reason that children consume antibiotics (Rothman et al. 2003; Rovers et al. 2004). It is also one of the most common reasons for dizziness and vertigo in children, although the mechanism remains obscure. Postural instability during otitis media with effusion (OME) results from pressure changes in the middle ear (Grace and Pfleiderer 1990). Others believe that serous labyrinthitis is responsible for the vestibular disturbances in children with chronic OME (Golz et al. 1998).

Pneumatic otoscopy is recommended as the primary and also the best diagnostic tool for clinicians to distinguish OME from acute OM. Tympanometry can be used to confirm the diagnosis of OME. In acute OM, ear pain is the most useful symptom (Rothman et al. 2003). Children with severe visual impairments should be considered more vulnerable to such OME sequelae as balance problems. Any balance problems or unexplained clumsiness with OME should be noted and documented (Takata et al. 2003; American Academy of Pediatrics, Clinical Practice Guideline 2004).

Most child patients with OME completely resolve after ventilation tube insertion (Crace and Pfleiderer 1990; Colz et al. 1998). Results of vestibular tests, such as spontaneous and positional nystagmus by ENG, Romberg, and past-pointing, also normalize after myringotomy and ventilation tube insertion (Koyuncu et al. 1999). Children with OME are more visually dependent for balance than healthy controls, and they also show increased postural sway during moving visual scene tests (Casselbrant et al. 1998). Because long-term sequelae, such as abnormal development of balance and vestibular function, may occur in children with recurrent or persistent OME, early intervention is important (Casselbrant et al. 2000; Cawron et al. 2004).

2.3.5 Vestibular neuronitis

The etiology of VN in children, as in adults, is unclear. However, about half of the children with VN have had a preceding upper respiratory tract infection (Dix and Hallpike 1952; Tahara et al. 1993; Taborelli et al. 2000). MRI with high-dose gadolinium showed enhance-

ment of the vestibular nerve during acute VN in two adults, supporting a viral and inflammatory cause for some patients with VN (Karlberg et al. 2004).

Children with VN typically recover within 2-4 weeks, and their prognosis is better than that observed in adults (Shirabe 1988). Vestibulo-ocular reflex defects also recover better than in adult patients with VN, indicating faster recovery of nystagmus and attenuated caloric responses (Tahera et al. 2000). VN was originally described by Hallpike in 1949. VN was found to chiefly affect the age group of 30-50 years, with no gender preference. Five percent of their patients with VN were under 20 years of age. They reported no cochlear signs or symptoms and otoscopy was normal. The disorder appeared to be aggravated by head movements of all kinds. The course of the disease was stated to be benign (Dix and Hallpike 1952). Typical clinical signs and symptoms in patients with VN comprise horizontal rotational nystagmus, which is aggravated by head motion, difficulties in standing and walking, and the tendency to veer towards the affected side. The symptoms of malaise, pallor, nausea, vomiting, and sweating are nearly always present. Vertigo develops quite suddenly, is severe for a few days, and gradually subsides over the course of a few weeks. Some patients may feel dizziness or imbalance for months after the onset of VN (Baloh 2003). No cranial nerve abnormalities are associated with VN. Headache and hearing loss are absent as well.

According to the original definition of VN, ENG should reveal pathology in a vestibular organ. The diagnosis of VN today is based on a typical clinical picture and exclusion of other possible causes of vertigo without putting too much emphasis on caloric responses, as these are limited to testing of horizontal semicircular canals.

2.3.6 Meniere's disease

Prospier Meniere already in an article in 1861 noted that children suffer from the same kinds of symptoms as adults; these symptoms were later to be known as MD. He described children with symptoms of a spinning feeling, pallor, vomiting, and a tendency to fall. After two or three vertigo attacks, the children sustained hearing loss. In his article, Meniere encouraged clinicians working with children to study this topic (Meniere 1861).

MD is about 100 times less frequent in children than in adults, with only 1% of affected patients being children (Hausler et al. 1987). According to Stahle et al. (1978), the overall MD incidence in Sweden was calculated to be 0.05%. In a Japanese study, the prevalence of MD in vertiginous children was 2.9% (Akagi et al. 2001). Meyerhoff et al. (1978) reported that 3% of Meniere patients were children. The prevalence of MD in a Finnish general population was 0.5% when the most recent criteria for MD were applied (American Academy of Otolaryngology - Head and Neck Surgery (AAO-HNS) 1995; Havia et al. 2005); however, no children under 12 years of age were included in this study.

Criteria for definite diagnosis of MD has been defined by the AAO-HNS in 1995 as follows: two or more definitive spontaneous episodes of vertigo lasting 20 minutes or longer, tinnitus or aural fullness in the treated ear, hearing loss on at least one occasion in audiometry, with other causes excluded. Other diseases to be excluded are perilymphatic fistula (PLF), posttraumatic vertigo, vestibular schwannoma, neuroborreliosis, and other CNS causes, such as epilepsy-related vertigo, MAD, vascular malformations, tumors, and multiple sclerosis.

In children, the clinical presentation of MD is not as typical as in adults, and the triad of vertigo, tinnitus, and sensorineural hearing loss is not as clear in very young children. Diagnosis of MD in children requires years of follow-up and careful examinations (Meyerhoff et al. 1978; Filipo and Barbara 1985; See et al. 2002) to exclude metabolic and inflammatory disorders and underlying acoustic or physical trauma (Meyerhoff et al. 1978).

2.3.7 Delayed endolymphatic hydrops

Delayed endolymphatic hydrops (DEH) is a disease entity that typically occurs in patients who have in childhood sustained a profound sensorineural hearing loss in one ear, usually from infection or head trauma, and after a long delay develop episodic vertigo in the same ear. This type of DEH is called ipsilateral DEH. The opposite ear can be affected and the patient has symptoms of fluctuating hearing loss with or without episodic vertigo. This type of DEH is called contralateral DEH (Schuknecht 1978). DEH always occurs in patients as a delayed manifestation of a pre-existing ear pathology (Hicks and Wright 1988). The delay between the discovery of deafness or profound hearing loss and the onset of vertigo episodes varied in one study from 1 year to 45 years, averaging of 26.8 years (Schuknecht 1978). In most patients, the delay is less than 25 years (Lambert 1985).

Causes of hearing loss include head trauma (Schuknecht 1978; Ylikoski et al. 1982; Hicks and Wright 1988), infections, such as labyrinthitis, meningitis, scarlet fever (Hicks and Wright 1988), mumps, influenza, and diphtheria, and unknown etiology. Acoustic trauma is reported to cause DEH (Ylikoski 1988). In one study, cytomegalovirus infection was the underlying cause of hearing loss and subsequent DEH (Huygen and Admiraal 1996). The etiology of DEH is also unknown. One theory is that simultaneously with the patient becoming symptomatic with vertigo the hydrops develops from an inflammatory process in the inner ear that has obstructed the endolymphatic duct or interfered with venous drainage from the endolymphatic sac (Lambert 1985).

The vertigo episodes in DEH are of the MD type. However, the DEH entity differs from MD in that way that the patients have histories of delayed vertigo after profound unilateral hearing loss. Ipsilateral DEH patients do not usually experience tinnitus or fluctuating hearing loss. The deaf ear on ENG testing shows reduced vestibular response (Lambert 1985). In the contralateral type of DEH, the opposite ear may have such symptoms as fullness, pressure, and tinnitus (Hicks and Wright 1988).

Vertigo in DEH is described as spinning with sudden onset and prolonged duration from one to several hours. The patient typically also has nausea and vomiting. In contralateral DEH, the patients develop fluctuating hearing loss in the opposite ear, also occasionally with vertigo (Schuknecht 1978). The hearing loss is usually discovered in childhood (Hicks and Wright 1988).

Surgical treatment for the ipsilateral type of DEH with disabling vertigo symptoms has given good results. Surgical techniques include labyrinthectomy and endolymphatic sac surgery (Schuknecht 1978; Hicks and Wright 1988). Younger patients, in particular, could be treated with endolymphatic sac surgery, and if it fails, vestibular nerve section can subsequently be performed (Hicks and Wright 1988). Contralateral DEH is treated conservatively

with medical therapy, but nondestructive surgery may also be considered if hearing loss or vestibular symptoms worsen (Hicks and Wright 1988).

2.3.8 Posttraumatic vertigo

The incidence of posttraumatic vertigo is not known, but its existence has been noted by Eviatar et al. (1986), who divided it into five major categories: labyrinthine concussion, whiplash syndrome, basilar artery migraine, vertiginous seizures, and nonspecific posttraumatic dizziness. This division facilitates setting of a diagnosis and initiating treatment (Eviatar et al. 1986).

In a study of adult patients, primary (vertigo occurring within 24 hours of trauma) neurotological disorders after head trauma included benign paroxysmal positional vertigo, labyrinthine concussion, PLF, and central vestibular disorder. Secondary disorders were diagnosed, on average, six weeks after trauma. The most important of these was DEH, others being cervicogenic vertigo and otolith disorders. Posttraumatic vertigo can manifest in different ways and can also have a late onset of symptoms, thus being challenging for otolaryngologists. No significant correlation was found between the mechanism of trauma and the type of neurotological disorder (Ernst et al. 2005).

The postconcussion syndrome has often been thought to be the reason for posttraumatic vertigo. However, PLF as a cause of posttraumatic vertigo should be considered, particularly in patients with persistent or intermittent vertigo or fluctuating hearing loss after head trauma (Jacobs et al. 1979).

Immediately after head trauma, almost half of children had spontaneous or positional nystagmus. Most children with posttraumatic vertigo recover within 6 months (Vartiainen et al. 1985). In adults, attacks of vertigo can develop a long time after the initial head trauma. The pathophysiologic explanation for vertigo attacks may be delayed hydrops in the inner ear (Ylikoski et al. 1982). Children with vertigo showed significantly disturbed body sway on posturography immediately after mild head trauma compared with age-matched controls who had been followed up for allergic problems but were otherwise healthy (Lahat et al. 1996). The onset of vertigo can occur a few weeks or even months after the initial trauma. The underlying disorder in posttraumatic vertigo, if identified, can be treated (Ernst et al. 2005).

2.3.9 Perilymphatic fistula

The incidence of PLF in children is unknown. It is an abnormal connection between the inner ear and usually the middle ear. The most common location for fistula is between the oval or the round membrane and the middle ear. This fistula allows perilymph to leak out of the inner ear, producing dizziness or vertigo and/or hearing loss (Fitzgerald 1995).

In PLF, vestibular disturbances have been proposed to be more frequent than hearing loss (Parnes and McCabe 1987). PLF should be suspected in patients with unilateral, most often sensorineural hearing loss (SNHL) seen in Ag after head injury, barotrauma, or direct force to the ear canal. Patients with significant and persistent dizziness after head injury may also have PLF (Parnes and McCabe 1987; Fitzgerald 1995). Diagnosis of PLF is challenging because the

symptoms can vary due to an intermittently open and closed PLF. Therefore, test results can differ from time to time (Fitzgerald 1995).

No specific criteria exist for diagnosing PLF preoperatively. The only certain way to diagnose PLF is by microscopic visual inspection during PLF repair (Weber et al. 2003). If no leak is visible from the inner ear, packing the round and oval windows did not affect vestibular status (Weber et al. 2003).

Vertigo may or may not be present in congenital PLF. Fitzgerald (1996) noted that younger individuals (teens and young adults) with pre-existing SNHL were more prone to developing an active PLF at some point in their lives than normal-hearing controls. SNHL is a treatable symptom.

Beta-2 transferrin test from perilymph confirms the existence of PLF in children and adults (Weber et al. 1995). Surgical repair of congenital PLF relieves the symptoms of vertigo in most children (Ruben and Yankelowitz 1989; Weber et al. 2003). With surgery, vestibular symptoms related to PLF can be treated more effectively than hearing loss (Parnes and McCabe 1987).

2.3.10 Psychological dizziness

The incidence of psychogenic vertigo or dizziness is unknown, but the disorder is often underestimated by clinicians who work with children. The most common complaints of patients with psychological disorders are headaches, followed by dizziness, and the most frequent disorder underlying these symptoms is depressive disorder. The most frequent psychosocial stress factor has been found to be school-related (Emiroglu et al. 2004). In adults, physical neurotologic conditions are known to trigger psychopathology, such as new anxiety or depressive disorders, as often as primary anxiety disorders cause dizziness (Staab and Ruckenstein 2003).

Neurologic symptoms, such as headache, vertigo, dizziness, and fainting, can be manifestations of a psychiatric disorder. Psychosocial stress factors, including school problems, familial dysfunction, parental psychopathology, and child sexual abuse, were associated with somatic symptoms. Psychological disorders, particularly depression, should be considered in evaluation of pediatric neurologic populations; early psychiatric consultation can prevent unnecessary and sometimes stressful physical examinations (Emiroglu et al. 2004).

2.3.11 Ocular disorders

Ophthalmologic problems, such as convergence insufficiency or latent strabismus with binocular vision, can cause vertigo. While the incidence is unknown, it is suspected to be quite high because of frequent computer use and television watching by children. Refractive errors, such as hyperopia, myopia, and astigmatism, may also underlie vertigo or dizziness (Anoh-Tanon et al. 2000).

Children diagnosed with ocular disorders are generally older than 6 years, and clinical manifestations are often accompanied by fatigue. Trigger factors are long exposure to computer or television screens. Most children recover with simple ophthalmic treatment. Anoh-Tanon et al. (2000) found that in 44% of children vertigo related to ocular disorders was asso-

ciated with headache, particularly in children with a familial or personal history of migraine. Moreover, vertiginous children with normal neurological findings and without obvious vestibulopathy after vestibular testing should undergo ophthalmologic consultation before more costly examinations, such as magnetic resonance imaging (MRI). Electro-oculography with saccade, smooth pursuit, vergence, and combined movements analysis is useful for diagnosis and treatment of children with vertigo (Bucci et al. 2004).

2.3.12 Neurological diseases associated with vertigo

When a child has a neurological disease, attention is usually paid to the disease itself and symptoms, such as dizziness, are neglected unless they are very disabling. Several CNS diseases may be accompanied by vertigo, dizziness, or ataxia. The prevalence of these disorders causing dizziness has not been established. Epilepsy, migraine (see above), multiple sclerosis, CNS tumors, CNS infections, transient ischemic attack, hydrocephalus, and malformations of the brain can all cause dizziness, vertigo, or ataxia (Blayney and Colman 1984; Eviatar 1994; Bower and Cotton 1995). In a study from a pediatric neurology clinic, a very high incidence of central vertigo was found (42/50), leaving only 8 children with a peripheral cause for vertigo. Central causes for vertigo or dizziness were seizures in most children (n=25), other less common causes being postmeningitic, posttraumatic, migrainous, and psychosomatic. Family history was positive for seizures in 13 children with seizures. Five of the children with seizures had had febrile seizures in infancy (Eviatar and Eviatar 1977).

If vertigo or dizziness is suspected to be of epileptic origin, the diagnosis should be confirmed with ictal EEG. A brain MRI should be performed to rule out tumors and structural anomalies. Unrelieved vertigo accompanied by cranial nerve deficits, pyramidal tract signs, or cerebellar abnormalities suggest a space-occupying lesion in the cerebellopontine angle or the posterior fossa, and imaging of the head is indicated. The MRI of the brain is helpful in ruling out demyelinating diseases, such as multiple sclerosis or neuroborreliosis, and degenerative CNS disorders that can manifest in vertigo or dizziness (Eviatar 1994).

2.3.13 Genetic syndromes associated with vertigo

Some genetic disorders are associated with vertigo. These include Pendred syndrome (PDS), Usher syndrome subtypes I and III, and coagulation factor C homology (COCH) gene mutations. Morphological anomalies of the inner ear, such as enlarged vestibular aqueduct (EVA), encountered in 15% of patients with PDS and in Mondini dysplasia may be caused by a mutation in the PDS gene that encodes the transmembrane protein Pendrin. The Pou Domain, class 3, transcription factor 4 (POU3F4) gene mutation, often causing the clinical signs of stapes fixation and gushers, may also manifest in abnormal vestibular signs and cause vertigo.

EVA is often part of syndrome disorders and usually appears in PDS syndrome. The clinical picture of EVA is typically fluctuating SNHL combined with episodic vertigo. Children with EVA can suffer from vertigo lasting minutes to hours (Oh et al. 2001). Because the clinical picture in some cases is very similar to PDS, EVA may be a milder variant of PDS rather than a different disorder (Stinckens et al. 2001). EVA was found to be frequent in the patho-

genesis of SNHL, especially when the onset of hearing loss was in infancy or childhood (Berrettini et al. 2005). Patients with PDS have been reported to have inner ear malformations, such as enlargement of the endolymphatic sac and duct and EVA, and less frequently Mondini dysplasia (Phelps et al. 1998). In Mondini dysplasia, the upper portions of cochlea are hypoplastic and form a common cavity. In a Japanese study, mutation in the PDS gene showed some correlation with the development of an enlarged endolymphatic sac and duct (Naganawa et al. 2004).

PDS is an autosomal recessive condition characterized by bilateral, sensorineural, and severe to profound hearing loss and by goiter with or without hypothyroidism. The gene mutated in PDS is located on chromosome 7 and encodes the pendrin protein. This mutated gene is mainly expressed in the thyroid gland, kidney, and inner ear. The PDS phenotype is highly variable regarding hearing loss and thyroid problems within a family. Environmental or other genetic factors therefore have an impact on the PDS phenotype (Napiontek et al. 2004).

The COCH gene, mutated in DFNA9, encodes cochlin, which is an extracellular protein expressed in spiral ligament and stroma underlying the vestibular sensory epithelium. The COCH gene causes autosomal dominant inherited hearing loss associated with vestibular dysfunction. The onset of deafness occurs between the second and fifth decades of life, with initial involvement of higher frequencies. Vestibular dysfunction is usually noted in patients with COCH mutations, and symptoms resembling those of MD, including vertigo, tinnitus, and aural fullness, were noted in 25% of patients (Tekin et al. 2001). The condition is rare. Cochlin protein acts as a master regulator that organizes the specific architecture of the extracellular matrix (ECM) in the cochlear and vestibular systems, and alterations in cochlin's ability to integrate into the ECM or to interact with specific ECM components may lead to DFNA9 deafness (Grabski et al. 2003). No mutations of the COCH gene were found in patients in hearing loss families without vestibular symptoms. MD patients also had no mutations of the COCH gene (Usami et al. 2003).

The Usher syndrome is an autosomal recessive disorder characterized by SNHL and retinitis pigmentosa. This syndrome is both clinically and genetically heterogeneous. Based on phenotypic variation, patients with Usher can be separated into three main types. In types I and III, patients have vestibular problems in addition to SNHL and retinitis pigmentosa. In type II, the vestibular function is intact (Kimbeling 2005). Noteworthy is that in the extension of the Framingham study the hearing loss was associated in genomic analysis with overlapping regions of known Usher genes. Thus far, it has not been documented whether expression of these genes also causes vertigo in children (DeStefano et al. 2003).

2.4 Diagnostic evaluation

Any medical doctor working with children in a hospital or in primary care is likely at some point to encounter a patient complaining of dizziness. Often by asking the right questions and conducting appropriate examinations, it is possible to distinguish between recoverable and progressive course of dizziness and to refer the child onward, to the right place, usually to either a neurotologist or a neurologist, for further investigations. The patient history is the most valuable diagnostic tool in this evaluation process. For example, in the diagnosis of BPVoC, a

family history of migraine is essential (Al-Twaijri and Shevell 2002). Also MAD in children can sometimes occur without the headache component, and a positive family history of migraine can lead to the correct diagnosis (Parker 1989).

2.4.1 Patient history

Many authors emphasize the importance of patient history in diagnostic evaluation of vertigo in children (Blayney and Colman 1984; Balkany and Finkel 1986; Britton and Block 1988; Eviatar 1994; Bower and Cotton 1995; Ravid et al. 2003). The history should be detailed and a family history included. Very young children have limited communication abilities and vocabulary to describe their symptoms but even so can often describe vertigo symptoms. The duration of dizziness and associated symptoms, such as nystagmus, ear symptoms, change in level of consciousness, headache, sensitivity to flashing lights, drooling, and blurred speech, as well as other events surrounding the attacks should also be ascertained. Past medical history should also include neo- and perinatal problems, such as sepsis and other infections treated with ototoxic drugs, because they can be an underlying cause for dizziness years later (Balkany and Finkel 1986; Eviatar 1994). Some conditions, e.g. BPVoC, can be diagnosed almost solely by taking an accurate history. An algorithmic and structured approach is recommended in evaluating a vertiginous child. To this end, a specific pediatric questionnaire that takes into account the child's age has been developed to make it easier for the physician to focus on relevant symptoms and avoid unnecessary examinations (Ravid et al. 2003, Table 2). After covering the variables in Table 2, a differential diagnosis with, for example, a computer-assisted algorithm can be assigned.

Table 2: Pediatric structured questionnaire (adapted from Ravid et al. 2003)

Age, years	< 5	> 5
Nature of symptoms	Yes	No
Vertigo		
Acute		
Paroxysmal		
Hearing loss		
Change of symptoms with head position		
Associated symptoms		
Headache		
Fever		
Vomiting		
Anxiety		
Depression		
Change in consciousness		
Head trauma		
Drugs		
Dizziness		
Chronic		
Continuous		
Family medical history		
Hearing loss		
Migraine		
Seizures		
	Normal	Abnormal
Neurologic examination		
Physical examination		

2.4.2 Office examinations

A primary care physician without sophisticated examination possibilities is also able to determine the otoneurological status of a child. Otologic examination should include complete ear, nose, and throat examination, and the ears should be investigated with pneumatic otoscopy. Tuning fork tests are helpful in differentiating between conductive and sensorineural hearing loss. Calorics can be done without recording devices and provide information on semicircular canal function. The Dix-Hallpike positioning maneuver, observation of Hennebert's sign, and observation of spontaneous or induced nystagmus with the help of Frenzel glasses should be included (Bower and Cotton 1995). Head shaking nystagmus can be helpful in evaluation of unilateral vestibular deficit (Fife et al. 2000). Other tests, such as Romberg, Unterberg, and hopping tests, reveal how good a patient's balance is. In neurologic evaluation, it is important to test all cranial nerves, visual fields, Romberg, Unterberg, finger-to-nose pointing, diadochokinesis, tandem gait, heel gait, toe gait, hopping, deep tendon reflexes, muscle tone, and muscle strength (Eviatar 1994).

2.4.3 Otoneurological tests

Children with vertigo can be evaluated using the same techniques used for adults, such as calorics and rotational chair testing, with a few minor adjustments to the methodology (Balkany and Finkel 1986; Levens 1988; Fife et al. 2000). However, the range of normal values in children varies more than in adults (Fife et al. 2000), and the data for children cannot be interpreted in terms of adult values. Children without vertigo have a higher incidence of spontaneous and positional nystagmus than adults (Levens 1988).

In the diagnostic work-up of childhood vertigo, important diagnostic tools were evaluation of hearing with Ag, tympanometry, and ENG (Britton and Block 1988; Bower and Cotton 1995). Children with persistent and severe ataxia or dizziness should undergo ENG, EEG, and imaging of the head (Blayney and Colman 1984).

Standard pure-tone audiometry (PTA) enables discrimination between sensorineural and conductive hearing loss. Ag reveals any asymmetry in hearing thresholds between ears. Hearing loss at specific frequencies can also be documented. Clear asymmetry, especially at higher frequencies, may suggest a diagnosis of vestibular schwannoma in a child with Neurofibromatosis II. With Ag, the progression of the disease can be followed and any fluctuations in hearing loss at different timepoints observed. This hearing fluctuation especially at the lower frequencies can be a sign of endolymphatic hydrops as in MD. PTA is a subjective test and thus demands cooperation, which can be insufficient in young children. Children from two to approximately five years of age can be motivated by play audiometry and younger ones by visual reinforced audiometry. Objective electrophysiologic tests, primarily auditory brainstem response (ABR), are used to estimate hearing sensitivity. These tests can be performed shortly after birth during an infant's natural sleep or under a mild sedation or general anesthesia. A study from Denmark noted that about 50% of children two years of age were able to establish thresholds in play audiometry at at least three frequencies and nearly 75% of children three years of age could establish six thresholds or more, and thus, do not necessarily need ABR (Nielsen and Olsen 1997).

In distinguishing OME from acute OM, pneumatic otoscopy is the primary diagnostic method, and tympanometry can be used to confirm the diagnosis of OME (American Academy of Pediatrics [AAP] 2004). The benefits of tympanometry over pneumatic otoscopy are the possibilities for documentation and increased diagnostic accuracy (AAP 2004).

ENG includes caloric irrigation, testing for spontaneous and positional nystagmus, smooth pursuit tracking, saccadic eye movements, and optokinetic nystagmus. Video-oculography is currently overtaking ENG because of its superior sensitivity. It differs from ENG in the method by which eye movements are recorded (Fife et al. 2000). ENG is a good method in distinguishing between central and peripheric vertigo (Eviatar 1994; Bower and Cotton 1995; Bakr and Saleh 2000). The majority of children respond to calorics and rotational testing within the first two months of age (Balkany and Finkel 1986; Fife et al. 2000).

A positional nystagmus when the head is in a particular position and without latency is indicative of a central lesion. Moreover, a pure vertical or pure torsional nystagmus is always caused by a central lesion (Salami et al. 2005). Calorics are helpful in documenting unilateral vestibular hypofunction, but for bilateral vestibular hypofunction, rotational chair testing is more specific. In children three years of age or younger, rotational testing is more convenient because the child can sit in the parent's lap during the test, and vertigo is less intense than in

caloric testing. In children five years of age or older, both caloric and rotational chair testing can usually be performed successfully (Fife et al. 2000). In another study, the authors emphasize the presence of nystagmus over decreased caloric response in diagnosis of peripheral vestibular disorders (Uneri and Turkdogan 2003). Casselbrant et al. (1998) demonstrated that children with OME have impaired postural responses and increased postural sway in response to moving visual surroundings compared with healthy controls.

In EEG, electrical activity is measured and registered with surface electrodes on standardized locations on the skull. This test gives particularly good information on cerebral cortical function. EEG wave forms depend largely on patients' age and state of alertness. Dizziness, loss of consciousness and altered behavior can be signs of epileptic seizures. A normal interictal EEG, seen in about half of epileptic patients, does not exclude the diagnosis of epilepsy, nor does an abnormal routine EEG necessarily establish it (Menkes 1985; Koskiniemi and Donner 1987). Thus, it is very important with ictal EEG to rule out true epileptic vertiginous seizures. The differential diagnosis between epilepsy syndromes and migraine is not always easy to make, as is the case in occipital lobe epilepsy with visual symptoms, dizziness, vomiting and headache (Sand 2003).

2.4.4 Laboratory tests

For most forms of vertigo, no specific screening tests are available. Eviatar (1994) recommends a complete blood count, serum evaluations for electrolytes, calcium, and magnesium, a glucose tolerance test, thyroid function tests (T3, T4, TSH), and immunoglobulin evaluations for all patients complaining of dizziness to exclude underlying causes of metabolic, endocrine, dysgammaglobulinemia, severe anemia, and sickle cell anemia disorders.

Others believe that routine metabolic screening tests are not helpful, but in suspicion of dizziness due to, for example, thyroid dysfunction, hypoglycemia, Addison's disease, and other metabolic or genetic disorders, laboratory tests can facilitate a diagnosis being set (Bower and Cotton 1995). If neuroborreliosis is suspected of being an underlying cause of vertigo, serum and cerebro-spinal fluid borrelia antibodies need to be examined. If a child on regular medication develops dizziness, medication should be considered as a cause of iatrogenic dizziness; especially some antiepileptics, such as carbamazepine, oxcarbazepine, and phenytoin, can produce dizziness (Eriksson et al. 2003).

2.4.5 Imaging studies

To rule out intracranial processes and also in cases of head trauma, imaging of the head is helpful (Britton and Block 1988; Eviatar 1994; Bower and Cotton 1995). However, imaging studies in children with headaches are of very limited value without clinical evidence of an underlying structural lesion. Brain imaging is indicated in children with headaches for whom a clinical history cannot be reliably obtained. Conversely, brain imaging should be avoided in cases of well-defined migraine because imaging is expensive and includes the additional risk associated with general anesthesia (Maytal et al. 1995). MRI of the head is of value when used specifically based on the symptoms and signs at presentation (Ravid et al. 2003). Children with

persistent ataxia or dizziness should undergo head imaging along with other relevant examinations (Blayney and Colman 1984). No systematic studies have been conducted concerning the value of imaging of the head in vertiginous children.

After head trauma, computerized tomography (CT) can reveal bone fractures. CT of the ears in chronic ear problems can show cholesteatoma, underlying pathology making the patient susceptible to PLF, or an inner ear anomaly, such as EVA or Mondini dysplasia, as an underlying cause for dizziness. Sinusitis can cause symptoms that a child describes as dizziness. If cold symptoms do not improve within 7-10 days, sinusitis should be seriously considered. In children, the sensitivity and specificity of sinus X-ray are poor. X-ray can, however, be helpful if an air-fluid level is seen. In cases of complicated sinusitis and when sinus surgery is being considered, a CT scan is necessary. CT should not be used for diagnostic purposes (Ramadan 2005). Acute respiratory infections cause mucosal edema in the paranasal sinuses of children. These mucosal abnormalities tend to resolve without antimicrobial treatment. Thus, clinicians should not make decisions on treatment based only on radiological findings (Kristo et al.2003).

3 Aims of the study

The aim of this project was to evaluate the prevalence and clinical characteristics of childhood vertigo.

Specific aims were as follows:

1. To determine the prevalence and characteristics of vertigo and balance problems in Finnish children aged 1-15 years. (I)

2. To evaluate –using a structured approach- the history and findings in vertiginous children as compared with a control group of healthy children. (II)

3. To assess the prevalence and characteristics of symptoms in vertiginous children visiting an ENT clinic. (III)

4. To determine the value of and indications for imaging of the head in vertiginous children. (IV)

4 Materials and methods

4.1 Subjects

In the prospective epidemiological study from the general population (I), altogether 1050 children in the HUCH area received a screening questionnaire and 938 (473 girls, 465 boys) returned it; thus, the response rate was 89%. The children were aged 1-15 years (mean 9.3 years). From the same child population, 30 vertiginous children with true episodes of vertigo of unknown etiology were invited to further examinations at the ENT clinic (II). True vertigo was defined as rotational or veering vertigo. Children with orthostatic hypotension and vertigo due to misuse of alcohol were excluded from the true vertigo study group. Of those invited, 24 subjects (15 girls, 9 boys) aged 2-16 years participated. In all, we studied 24 vertiginous children and 12 healthy age- and gender-matched controls. Their mean ages were 10.4 years and 10.3 years, respectively. Girls made up 62% of the study group and 58% of the control group. In Study III, we reviewed the medical records of all subjects with vertigo, altogether 119 children (63 girls, 56 boys) aged from 7 months to17 years (mean age 10.9 years at ENT clinic examination), who visited the ENT clinic between 2000 and 2004 with a primary complaint of dizziness or vertigo. In Study IV, we reviewed the medical records of 978 children who had an MRI or CT of the head or a CT of the ears in 2004. Of these, 87 (40 boys, 47 girls) had imaging of the head because of vertigo, and we studied them more carefully. We also more closely examined 23 vertiginous children (13 girls, 10 boys) with a new abnormal finding in images that was thought to be the underlying cause of vertigo. The Ethics Committee of the Department of Otorhinolaryngology, HUCH, approved the study protocols (I-IV).

4.2 Methods

In Study I, we collected data on balance problems and vertigo from children in three different schools and one child welfare clinic in the HUCH area. The simple screening questionnaire with mostly yes/no types of questions also focused on recurrent falls, difficulties in walking, clumsiness, peculiar behavior, and experienced fear or panic (Appendix in Study I). At the welfare clinic, the questionnaire with an information letter was given to 300 consecutive children visiting the clinic. At the three schools, we gave a questionnaire to every child in selected classes. The younger children filled in the questionnaires with their parents and the older children by themselves. The information on prevalence of vertigo and balance problems and other conditions was stored in a database for analysis.

In Study II, we examined 24 children with true vertigo from the subject pool of Study I at the ENT clinic. These children provided a detailed history, underwent otoneurologic and general examinations, and had Ag, ENG, and tympanometry done if the cooperation was sufficient. A group of 12 healthy controls underwent the same protocol. All data were

stored in a computer and analyzed by ENT doctors experienced in examining vertigo patients. We aimed to assign a diagnosis to all children.

In Study III, we collected data from the medical records of 119 vertiginous children who had visited the ENT clinic. Data included the nature of vertigo symptoms (acute or chronic, paroxysmal or continuous, attack severity, number and duration of attacks), provoking factors, ear symptoms (aural fullness, tinnitus, pain, infections, ear operations, hearing loss), other associated symptoms, examinations done thus far due to vertigo, past medical history, and any previous consultations with other medical specialists. The data were stored in the database and analyzed by ENT doctors experienced in treating vertigo patients, and all children were assigned a diagnosis.

In Study IV, we reviewed the medical files, including imaging reports of every child who had had an MRI or CT of the head or a CT of the ears in 2004. Out of 978 children, we analyzed closely the images and medical records of 87 children who had undergone imaging of the head due to vertigo, or dizziness during the study year. We paid attention to patient's age, gender, indication for imaging, possible traumas, neurological symptoms, radiological findings, and other possible diseases. Images with deviant findings were reviewed by an experienced neuroradiologist from the Department of Radiology at HUCH. All data were stored in the database, allowing us to determine the clinical signs warranting imaging of the head in vertiginous children.

4.3 Statistical analysis

For statistical analysis, we used SPSS statistical program version 10.0. Frequencies, means, ranges, and standard deviations were calculated for most of the variables in Studies I-IV.

5 Results

5.1 An epidemiological study on childhood vertigo (I)

The objective was to determine the prevalence and characteristics of vertigo and balance problems in Finnish children aged 1-15 years.

Seventy-five children (8%) had experienced vertigo, with prevalence being cumulative such that older children had experienced more vertigo in their lives than younger children. The majority of the children (48%, n=36) had 1-2 attacks a year. Of the vertiginous children, six (8%) had frequent attacks, at least once a week. The vertigo attacks varied in duration from 1-15 s (35%, n=26) to more than 4 hours (3%, n=2). Vertigo attacks interfered with normal activities in 23% (n=17) of vertiginous children.

Recurrent falls were experienced by 1% of the children and difficulties in walking by 2%. Clumsiness was most common in children aged 1-5 years; altogether 3% of the children had been exceptionally clumsy. Peculiar behavior was reported by 2% and occasional fear or panic by 3%. A possible provoking factor or reason for vertigo was mentioned in 69% of vertiginous children's questionnaires.

5.2 Diagnostic evaluation of vertiginous children (II)

In Study II, we aimed to evaluate history and findings in vertiginous children as compared with a control group of healthy children.

In the vertiginous group, there were significantly more head traumas than in controls ($p<0.05$). The two groups did not differ significantly in gestational age, birth weight, number of neonatal or other serious infections, or travel sickness.

In the children with true episodes of vertigo, the most frequent forms were BPVoC (5 children), MAD (4), and vertigo related to ear infections (4). In 2 children, no definitive diagnosis was established. Less frequent forms of vertigo were vertigo related to a fast period of growth (2 children), hypoglycemia (1), epilepsy-related vertigo (1), stress-provoked dizziness (2), posttraumatic vertigo (2), and psychological disorders (1). The characteristics of the children with BPVoC and MAD are presented in Table 3. The mean age of children at the onset of symptoms and at the examination were in BPVoC 6 and 9 years and in MAD 9 and 13 years, respectively. Attack frequency was higher but attack duration shorter in BPVoC children than in MAD children.

Table 3: Characteristics of children with BPVoC and MAD

	Gender	AO	AE	Attack frequency	Attack duration	Provoking factor
BPVoC	M	5	9	weekly	30 seconds	not known
BPVoC	F	4	7	daily	few seconds	not known
BPVoC	F	2	6	twice a month	few minutes	not known
BPVoC	F	8	13	weekly	10 seconds	not known
BPVoC	F	10	11	weekly	less than one minute	hunger
mean		6	9			
MAD	M	12	16	monthly	few minutes	lack of sleep, stress, missing a meal
MAD	M	9	11	weekly	5 minutes	excitement, stress
MAD	F	8	11	3 times a year	10 minutes to one hour	tiredness, physical strain, thirst
MAD	F	7	12	once a year	5-30 minutes	travelling in a car, stress, missing a meal
mean		9	13			

AO = age of onset
AE = age of examination
M = male
F = female
BPVoC = benign paroxysmal vertigo of childhood
MAD = migraine-associated dizziness

We also calculated the prevalence of vertigo and dizziness in all children based on questionnaires and examinations at the clinic. The most common cause of vertigo and dizziness was orthostatic hypotension, followed by hypoglycemia, BPVoC, OM-related dizziness, tiredness, and MAD.

Based on our results, the otoneurological examinations did not differ between the study group and the control group. Head traumas and headaches were more common in vertiginous children than in controls.

5.3 Vertigo and imbalance in children visiting the ear, nose, and throat clinic (III)

In Study III, the objective was to evaluate the prevalence and characteristics of symptoms in vertiginous children visiting an ENT clinic.

The diagnoses are shown in Table 4. Most children (n=92, 77%) had normal hearing in Ag, with no asymmetry and hearing thresholds equal or better than 20 dB hearing level, 22 (18%) had abnormal Ag, and Ag was unavailable for 5 children (4%). Two girls with a final diagnosis of MD had sensorineural and unilateral hearing loss as well as hearing fluctuation documented in Ag.

Table 4: Diagnosis of 119 children with vertigo

DIAGNOSIS	Number of children
Benign paroxysmal vertigo	23
Migraine-associated dizziness	17
Vestibular neuronitis	14
Otitis media-related dizziness	12
Psychogenic vertigo	6
Vestibulopathy (unknown)	6
Posttraumatic vertigo	6
Inner ear irritation, sudden deafness	4
Labyrinthine hydrops	4
Tension neck	4
Orthostatic hypotension	4
Epilepsy-related vertigo	3
Meniere´s disease	2
Chronic cholesteatoma and surgery	2
Mal de barquement	1
Benign paroxysmal positional vertigo	1
Autoimmune thyreoiditis, with hypothyreosis	1
Insulin shock-related vertigo	1
Sinusitis-related vertigo	1
Chiari I malformation	1
Ataxia (genetic)	1
Postoperatice vertigo (after astrosytoma operation)	1
CATCH 22 syndrome	1
Ophthalmic vertigo	1
Otitis media-related vertigo and migraine-associated dizziness	1
Mononucleosis	1
TOTAL	**119**

CATCH 22, cardiac defects, abnormal facies, thymic hypoplasia, cleft palate, and hypocalcemia

ENG was performed or attempted in 79 children. Six children did not complete the test because of insufficient cooperation. There were unilaterally reduced vestibular responses (side difference greater than 25%) in 12 patients with a diagnosis of sudden deafness, MD, posttraumatic vertigo, cholesteatoma, or VN. None had bilateral vestibular function loss.

Ninety of the 119 children (76%) were examined at the Department of Child Neurology, Hospital for Children and Adolescents, HUCH; 14 (16%) had deviant neurological findings. Imaging of the head was done for 71 children (60%). It was normal in 64 children (90%) and abnormal in 7 (10%). Two of these abnormalities were posttraumatic fractures, one postoperative condition after brain tumor operation, one anomaly in the semicircular canals, one Chiari I malformation, one unilateral labyrinthitis, and one nonspecific postbleeding sign. An ophthalmologic examination was carried out in 23 children who, based on history, were thought to have eye-related dizziness, but only one child's vertigo was purely ophthalmologic in origin.

5.4 Value of imaging studies in vertiginous children (IV)

The aim of this study was to determine indications for imaging of the head in vertiginous children.

Their ages ranged from 6 months to 16 years (mean 8.1 years). Of these vertiginous children, 53 (61%) underwent MRI, 24 (28%) CT, and 10(11%) had both examinations done. Abnormal findings were present in 37 children's images; 14 had previously confirmed pathological findings that were unchanged and did not explain their new onset of vertigo, and 23 had a new abnormal finding explaining the vertigo symptoms. Of the 23 children with vertigo and a new finding in images, 19 (83%) had other neurological signs as well. Four children with a new finding in images had no neurological deficits, but 3 of them had intense headaches and 1 a temporal bone fracture after head trauma. Of the 23 vertiginous children with a new finding in images, 17 had MRI, 1 with a cerebro-spinal fluid shunt problem had CT, and 5 had both examinations done. There were 33 vertiginous children (38%) with deviant neurological signs; 19 (58%) had an abnormal image, 2 (6%) had no changes from previous images, and 12 (36%) had a normal imaging study. All children with an obvious pathological finding in images (e.g. brain tumors, multiple sclerosis) had cranial nerve deficits or intense headaches. Multiple sclerosis lesions and acute disseminated encephalomyelitis (ADEM) signals can be difficult to distinguish in head MRI; however, the final diagnosis is always made based on clinical findings (Figures 1 and 2).

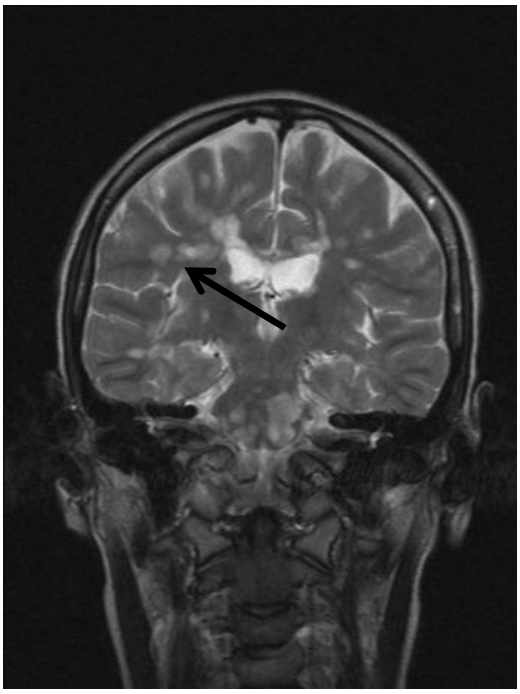

Figure 1: T2 MRI of a 15-year-old girl with multiple sclerosis lesions.

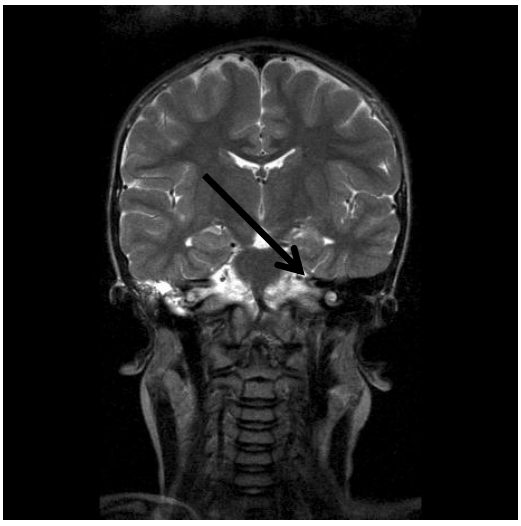

Figure 2: T2 MRI of a 4-year-old boy with acute disseminated encephalomyelitis signals.

6 Discussion

Studies I and II focused on gathering information on vertigo from a general child population. We found that 8% of the children had experienced vertigo or dizziness. Due to severe vertigo, 23% of these children had had to stop their activity. A thorough history should be obtained from vertiginous children; especially previous head traumas and occurrence of headache gave valuable information and were more frequent in vertiginous children than in controls. The predominant types of vertigo were OM-related vertigo, MAD, and BPVoC. We reviewed patient data of vertiginous children who visited an ENT clinic during 2000-2004. The most common diagnoses were BPVoC, MAD, VN, and OM-related vertigo. Valuable diagnostic tools in achieving a diagnosis were medical and family histories, otoneurologic examination, ENG, and Ag. We reviewed the medical papers of children who had imaging done due to vertigo. Head imaging in vertiginous children appears to be helpful only when the child has neurological deficits, or sustained head trauma along with vertigo. If vertigo is the only symptom, imaging studies of the head are unlikely to aid in diagnostic work-up.

As in adults, the history and otoneurologic examination are the basis for the majority of diagnoses. In children a proper history and a description of symptoms may be incomplete, due to the lack of vocabulary. In younger children and infants, parental observation constitutes the history.

Balance problems are not uncommon in children and can limit daily activities. In Study I, we attempted to differentiate between normal and abnormal clumsiness in very young children. We did not aim to diagnose the children, but requested their opinion on provoking factors or the cause of vertigo in the questionnaire. This information was identified by 69% of the children or their parents. None of the children reported trauma-related vertigo, perhaps due to the long lag time between the trauma and vertigo onset, making the correlation difficult to recognize. Posttraumatic vertigo has been cited as a frequent cause of vertigo in some studies (Eviatar and Eviatar 1977; Bower and Cotton 1995; D'Agostino et al. 1997; Choung et al. 2003).

The response rate was 89%. The rate would have been somewhat higher had some children, especially teenage boys, not responded jokingly to their screening questionnaires. We excluded any answers that were unclear or frivolous. It is noteworthy that the study focused on the population located in southern Finland. Hazards, accidents, and disease profiles may be different in non urban areas or in populations of other genetic backgrounds.

As far as we know, there are no earlier studies on prevalence of vertigo in children from the general population. Previous studies have concentrated either on school-aged children only (Abu-Arafeh and Russell 1995) or on children who were patients in ENT or neurological clinics, thus not revealing the true prevalence of vertigo in children (Blayney and Colman 1984; Bower and Cotton 1995; Choung et al. 2002; Ravid et al. 2003). When children with vertigo are referred to an ENT clinic, the reason for vertigo is considered to be peripheral, while children with suspected central problems, MAD, and other paroxysmal vertigo attacks are referred to a neurological clinic.

In Study II, we aimed to diagnose the children with vertigo of unknown etiology. In conjunction with the detailed history-taking, Ag and ENG yielded the most information. The number of children in both groups was small, and thus, definitive conclusions could not be drawn. We had problems in motivating the children to participate in studies at the clinic, as many of them no longer had vertigo symptoms and the examinations at the ENT clinic were time-consuming. The controls were also difficult to recruit. Most of the children cooperated well. Children with true vertigo were selected to undergo further examinations. Based on the screening questionnaires, it was sometimes difficult to identify the children with true vertigo and therefore suitable for the study and in need of further examinations. Children reporting, for example, orthostatic hypotension or dizziness after alcohol drinking were, however, quite easy to exclude from the study group.

In Study III, we reviewed medical records and determined medical characteristics of all children who had visited the Helsinki University ENT clinic because of vertigo. The major challenge here was to obtain a proper patient history from often poorly documented medical records. Children with vertigo made up 0.7% of the child population visiting the ENT clinic during this 5-year period. According to an epidemiological study, there should have been many more children with vertigo (Russell and Abu-Arafeh 1999). Our diagnosis results were in line with earlier studies done in ENT and neurology clinics (Blayney and Colman 1984; Eviatar 1994; Bower and Cotton 1995; Weisleder and Fife 2001; Ravid et al. 2003). The most common forms of vertigo were OM-related vertigo, BPVoC, MAD, and VN. We had two children with MD (1.7%). In the literature, MD prevalence in studies of vertiginous children has varied from 1.5% to 2.9% (Hausler et al. 1987; Akagi et al. 2001). The most common forms of vertigo were peripheral. This is as expected since the children were seen in an ENT clinic. Close cooperation between specialists is essential in establishing a diagnosis. Most of children (76%) had first visited a pediatrician or a child neurologist and had already undergone neurologic evaluations. When the underlying reason for vertigo was thought to be ear-related or unclear, the child was referred to an ENT clinic for further evaluation.

In Study IV, we reviewed all medical papers of children who had undergone head imaging or CT of the ears during the study year. Abnormal radiological findings were found in 37 out of 87 children who had imaging done due to vertigo or dizziness. There were 23 children with a new abnormal finding. In their history and at examination, 19 patients had concomitant neurological signs or deficits, 3 intense headaches, and 1 a previous head trauma. Children in this study had more central causes of vertigo, reflected in their first being referred to the Hospital for Children and Adolescents, not to an ENT clinic. The most common abnormalities in images were brain tumors, CNS infections, and multiple sclerosis lesions. As far as we know, there are no earlier studies concerning the value of head imaging in vertiginous children. This study was important to define the signs and symptoms of children who need imaging of the head. This information will help to diminish unnecessary and expensive examinations, and young children can avoid unnecessary general anesthesia.

Of the 23 children with a deviant image, 17 had undergone MRI, 1 with shunt problems had CT, and 5 had both MRI and CT. Radiation doses of CT scans may be harmful to children, and therefore, unnecessary head CT should be avoided (Khursheed et al. 2002). The first choice of imaging should always be MRI when available. Nevertheless,

CT of the head is preferred after head trauma or in children with shunt problems. With recent developments, MRI has become a very sensitive method of neuroimaging but can also reveal clinically insignificant findings. To counteract the high sensitivity of MRI, clinical findings should always be related to imaging results.

In Study III, the prevalence of psychogenic vertigo was 5%, which is relatively high. We are planning a further study on psychogenic comorbidity in vertiginous children. We also intend to establish a structured questionnaire for pediatric vertigo patients that can be stored electronically. This is anticipated to improve the quality of patient medical records, particularly as they pertain to patient histories. Although vertigo in children is not common, it is seen frequently enough to warrant an adequate understanding by all otologists and child neurologists.

7 Conclusions

Based on Studies I-IV, the following conclusions were drawn:

1. Balance problems or vertigo are not rare in children. Of the Finnish capital area population, 8% of children had at some point experienced vertigo, dizziness, or balance problems. Of these, 23% had sufficiently severe vertigo to prevent continuation of their activity.

2. The structured data collection approach eased the evaluation of vertiginous children. Otoneurological examinations did not differ between the study group and the controls. More head traumas and headaches were observed in vertiginous children than in healthy controls.

3. Vertiginous children comprised 0.7% of children visiting an ENT clinic during the 5-year period. The most common diagnoses were BPVoC, MAD, VN and OM-related vertigo. In the diagnostic process, the most valuable tools were patient history, otoneurological examination, ENG, and Ag.

4. Imaging of the head with MRI or CT is indicated for those vertiginous children with either neurological deficits or persistent headache, or after head trauma. If vertigo is the only symptom, imaging studies are not likely to be helpful in setting a diagnosis.

8 Acknowledgements

This work was conducted at the Department of Otorhinolaryngology, Helsinki University Central Hospital, in 2000-2006.

Professors Pekka Karma and Jukka Ylikoski and Docent Hans Ramsay are thanked for providing me with the opportunity to carry out this work.

My deep gratitude is due to my supervisor, Docent Erna Kentala, of the Department of Otorhinolaryngology for her constructive guidance and support. Over the years, we have had many frustrating moments, especially during Erna's two-year visit to USA, but also lots of joy. Although Erna has recently been extremely busy running the Audiology Department, she has found time to answer my questions. Her knowledge of computers is particularly impressive.

I am also sincerely grateful to Professor Ilmari Pyykkö, my other supervisor, for his support, enthusiasm, and constructive advice. He appears to effortlessly correct text, making it more rational.

I warmly thank my coauthors neuroradiologist Leena Valanne and otologist Sylvette Wiener-Vacher from France.

Docent Juha-Pekka Vasama and Docent Tuula Lönnqvist, the referees of this thesis, are thanked for constructive criticism and valuable comments on the manuscript. Editing language of this manuscript by Carol Ann Pelli is greatly appreciated.

During my years of specialization at the clinic I have had the opportunity to get to know many nice colleagues; Mervi Kanerva, Karin Blomgren, Mari Havia, Johanna Nokso-Koivisto, Mari Hero, and Satu Passinen, in particular, have lightened the days at the clinic. Karin Blomgren and Mari Havia are also acknowledged for their substantial contribution to the study. Specialists and all fellow residents at the Department of Otorhinolaryngology are thanked for pleasant collaboration. Research with Docent Antti Mäkitie has been a pleasure.

Working as a clinical teacher has been a great and challenging opportunity. We have a good working atmosphere at the Teaching Department, for which I thank Docent Petri Mattila, Karin Blomgren, and Antti Aarnisalo, for the period of time that we worked there together also Kimmo Leskinen and Teemu Kinnari. My sincere thanks are owed to Mirja Haapanen, the Head Nurse of the Teaching Department, for her generous help with practical problems and for being so easy to approach. Also Taimi Nordström is thanked.

I thank Irma Hytönen for technical help in data collection.

I also thank the auditory unit for performing the audiological and otoneurological examinations. Especially Raija Lehtimäki is acknowledged for her extra work in the evenings.

My heartfelt gratitude goes to my beloved husband, Janne, for his ongoing love and support. Arttu, Jaakko, Fanny, and Sandra, our four children, are my sunshine and the joy of my life. I am deeply grateful to my mother Tuulikki and my father Risto for providing me with a solid foundation and unconditional love. My sisters Sari, Meri, Katri, and Maria and my brother Larri are wonderful friends and great fun to be with. I am particularly grateful to my twin sister Sari for being my best friend. I am always able to share my joys and worries with her. My wise and lovely grandmother, Sirkka, I thank for love and sup-

port. I also thank my other, now deceased, grandmother and grandfather, a PhD himself, for love and support. I know that they would have been very proud of me today. Special thanks go to my parents-in-law Raili and Osmo, who are always ready to extend us a helping hand.

I also thank my many friends outside this clinic with whom I have shared countless joyful moments for reminding me that there is life beyond otorhinolaryngology.

Financial support from the Päivikki and Sakari Sohlberg Foundation, Otolaryngology Foundation, and an EVO grant is gratefully acknowledged.

Helsinki, August 2006

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